Modern Concepts of Cardiovascular Disease

Published monthly by the AMERICAN HEART ASSOCIATION

1775 BROADWAY AT 58TH STREET, NEW YORK, N. Y.

DR. THOMAS J. DRY, Rochester, *Editor*

DR. HOWARD B. BURCHELL, Rochester, *Associate Editor*

VOL. XVII NOVEMBER, 1948 No. 11

PHYSIOLOGIC MEASUREMENTS IN CARDIAC MALFORMATIONS

In the past year two articles[1,2] have appeared in this publication on the application of certain physiologic data to the diagnosis and interpretation of symptoms and signs of congenital heart disease. The clinical picture associated with the various anatomic defects has been more clearly drawn and accuracy of the clinical diagnosis of the rarer types of malformation has been increasing. Recently there has been a trend to regard some anomalies as much more common than previously indicated by the pathologic reviews; for example, pulmonary stenosis without associated ventricular septal defect, and tricuspid atresia.

A short review of varied physiologic measurements in cardiovascular malformations with discussion of the effect of treatment on some types may be of help in the understanding of the circulatory dynamics. It should be emphasized that the great advances made through special technics, as exemplified by catheterization of the heart, have not outmoded the long-established methods of diagnosis, and indeed in the majority of instances the recommendation of surgical procedures for correction or alleviation of cardiac malformations may be made on clinical grounds alone[3,4]. Such facts are important in that involved investigations associated with cardiac catheterization must be considered to have potential risk through the development of serious arrhythmias or the complications of endocardial injury. In our own moderate experience with cardiac catheterizations, we have regarded the appearance and persistence of a marked extrasystolic arrhythmia as an indication that the procedure should be terminated.

Intracardiac Malformations without Cyanosis

Investigations on patients with atrial septal defects have revealed that the basic disturbance in circulatory dynamics is due to atrial shunts from left to right of as much as 16 liters of blood per minute and an associated great increase in the output of the right ventricle. In the pulmonary artery the pressure has shown no direct correlation with the volume of flow and the pressure as a rule has been only moderately elevated. Great increases in pulmonary arterial pressure have been associated with little flow rather than the reverse. It has been found that interatrial mixing of blood, with the resultant slight but significant drop in oxygen saturation of arterial blood may occur in the presence of cardiac compensation and a large left to right shunt.

The cause for the pressure gradient from the left to the right atrium has been a debatable point. Recent investigations of Dexter and co-workers in which the pulmonary capillary pressure at the venous end has been measured point to the explanation that considerable difference in pressure in the venae cavae and the pulmonary veins would cause the pressure gradient and flow from the left to the right atrium.

In ventricular septal defects, it is unknown to what extent large defects may present themselves, first as uncomplicated conditions without cyanosis, and later some degree of cyanosis and polycythemia develop so that the clinical picture of the Eisenmenger complex is present. It is known, however, that marked pulmonary hypertension and the shunting of a large quantity of blood from the left to the right ventricle may occur with normal oxygen saturation of arterial blood.

One of the most interesting demonstrations of the value of recording pressures through the catheter, introduced into the heart, has been in cases of pulmonary stenosis in which the systolic pressures between the right ventricle and pulmonary artery differ greatly. The anatomic diagnosis may be particularly simple when pulmonary stenosis is not associated with a septal defect. When pulmonary stenosis is of the pure type, it is caused by a conical valvular stricture, and it has seemed easier to catheterize the pulmonary artery than when the right ventricular outflow tract is stenosed as in most cases of tetralogy of Fallot.

Patent Ductus Arteriosus

In general, the studies by cardiac catheterization in patent ductus arteriosus have confirmed the results of Eppinger and co-workers obtained by direct sampling of blood from the pulmonary artery during surgery. In the average case of patent ductus arteriosus flow in the ductus may be expected to be half or more of the output of the left ventricle. The pulmonary pressures, although usually only in the upper range of normal, on occasion have been so greatly increased as to constitute real pulmonary hypertension. Taylor, in the laboratories at the Mayo Clinic, has measured pressures in the pulmonary and systemic arteries during surgery for ligation of patent ductus arteriosus. He has found immediate rises in the systolic and diastolic pressures in the radial artery and slight decreases in pulmonary artery on closure of the ductus. The resultant changes in the systemic pressures are similar to those obtained on the closure of an arteriovenous fistula except for the absence of a bradycardiac response. The absence of such a response could presumably be related to the anesthesia. It is of interest that in those cases in which the flow in the ductus was measured, the amount of flow correlated linearly with the product of the radius of the ductus squared and the difference in the mean pressures in the pulmonary and radial arteries. The flow in the ductus did not correlate with either the diameter of the ductus or the difference in pressure alone. The accurate recording of pressures through a catheter is technically difficult but the pressures obtained during preoperative catheterization and during surgery by direct puncture of the pulmonary artery give an excellent correlation of the high and low (systolic and diastolic) values. It has been suggested that an abnormal increase in pulse pressure during and after exercise might be an aid in the diagnosis of patent ductus arteriosus. In preliminary investigations in which patients who have a patent ductus arteriosus have exercised and pressures have been recorded by means of an indwelling needle in the radial artery, specific or diagnostic responses have not been obtained.

Although cardiac catheterization is not indicated in the usual case of patent ductus arteriosus, it is indicated when associated defects, of the atrial or ventricular septum in particular, are suspected. In two such complicated cases of ductus arteriosus in which the various shunts have been studied and surgical therapy consisting only of closure of the duc-

tus has been carried out, it has been of considerable value to watch the oxygen saturation of arterial blood by the photo-electric oximeter throughout the operation and to have noted no decrease in saturation. The ultimate value of simple closure of the ductus in such cases is unknown as yet.

The rare syndrome of a patent ductus arteriosus with coarctation of the aorta or an anomalous vascular ring may sometimes present a confusing picture and the physiologic laboratory may contribute greatly to the understanding of the situation by supplying data on the exact pressure and oxygen content of the blood in both radial arteries and a femoral artery.

Congenital Heart Disease with Cyanosis

Extensive physiologic data on the cyanosis-producing types of cardiac malformation, particularly as obtained by cardiac catheterization and by ventilation studies with their interpretation have been discussed by Bing and associates[2]. In addition simple measurements of circulation times as discussed by Hitzig[5] may give evidence, usually of a corroborative nature, of a venous-arterial shunt. In our laboratories the development of improved oximeters and a technic for continuous sampling of arterial blood by an indwelling arterial needle have afforded the opportunity of studying in some detail the decrease in oxygen saturation of arterial blood in the course of exercise that is so characteristic of these cyanotic states. It has seemed remarkable to us that a few patients still will volunteer to exercise when the oxygen saturation of arterial blood is less than 30 per cent and still will be alert and conscious when it is less than 20 per cent.

If the oxygen content of mixed venous blood from the right atrium (obtained by cardiac catheterization), of arterial blood and of pulmonary venous blood (which could justifiably be assumed to be normal in the majority of cases) is known, the relative amount of venous blood by-passing the lungs may be readily calculated.

Such calculations have shown that the venous shunt in a variety of anatomic conditions has remained remarkably constant for each patient under varying environmental conditions of stress. Such observations support the thesis that the magnitude of the venous shunt is determined mechanically by the structural defect. As a corollary, these same studies indicate that the decrease in oxygen saturation of arterial blood on exercise is caused by a decrease in the oxygen content of venous blood shunted rather than an increase in the magnitude of the shunt. There is an increased desaturation of the blood in the capillaries for which any increase in cardiac output cannot compensate. The title of Blalock and Taussig's original article "Surgical Treatment of Malformations of the Heart in Which There is Pulmonary Stenosis or Pulmonary Atresia" stressed the basic mechanical nature of the defect for which surgical therapy was performed. Laboratory investigations have since given further physiologic support to their concept of the problem and a method for its alleviation.

Repeated studies for more than a year in our laboratory on one patient who had a well-compensated congenital defect with cyanosis has demonstrated a marked uniformity in oxygen saturation of arterial blood during rest. This patient presented also a consistent pattern of response in oxygen saturations of arterial blood on exercise and when given 100 per cent oxygen. As we have used the decrease in oxygen saturation of arterial blood with exercise as a probable reliable index of the degree of inadequacy of the pulmonary blood flow, it was of value to know this likelihood of reproducibility in a patient under relatively the same internal and external state.

The exact place that cardiac catheterization will eventually occupy in the diagnosis of the congenital cardiac malformations with cyanosis will be determined by the publication and evaluation of the extensive studies taking place in many medical centers. In our own small experience, the indications usually pertain to the question of the presence or absence of hypertension in the pulmonary artery. Thus the patient for whom catheterization is most likely to be recommended is one whose pulmonary artery is demonstrable in the routine roentgenogram of the thorax.

Coarctation of the Aorta

The cure of hypertension of the upper part of the body associated with congenital stricture of the aortic arch has been discussed in this publication by Gross[6]. The surgical cure of the condition has left many problems concerning the dynamics of the circulation unanswered. Although gratifying results are to be expected when the coarctation can be removed and end-to-end anastomosis accomplished, Clagett's experience at the Mayo Clinic has been that the exact clinical result cannot be predicted from the technical perfection of the reconstructed aorta. The persistence of moderate hypertension of the upper part of the body after operation is not uncommon but often the hypertension improves constantly for many months. As auscultatory determinations of blood pressure in the arm are subject to nonspecific influences, it was thought that the completeness of the cure of the coarctation, meaning the restitution of the normal state, might be best judged by the change toward normal in the pulse in the femoral artery and the ratio of systolic and diastolic pressures in the femoral artery to those in the radial artery. For this purpose Brown and Wood recorded the intra-arterial pressures of the radial and femoral arteries before and after operation by technics developed in the laboratories of the Mayo Clinic. As yet the record of the femoral pressure has not had a perfectly normal contour nor have the ratios been completely normal after operation. The interesting phenomena of continued improvement (or decrease) in pressure in the radial artery months after surgery have been observed without change in the ratio of femoral to radial pressure on the one hand. On the other hand continued improvement of the ratio of the pressures in the femoral artery to the radial artery has been observed without continued decrease in the moderate hypertension in the upper part of the body. It is hoped that such studies carried out over some years on postoperative patients will make possible an assay of the efficacy of surgical therapy and will indicate the proper age range for such therapy. Our studies have generally pointed to the conclusion that anastomosis of the left subclavian artery to the thoracic aorta, for the correction of coarctation has been an unsatisfactory procedure. Hallenbeck has recorded marked increases in pressure in the proximal portion of the aorta as judged by the direct pressures in the right radial artery when the left subclavian artery is clamped as a preliminary part of the reconstruction, and failure to decrease to less than the original values on completion of the reconstruction even though the lumen at the site of anastomosis of the left subclavian artery to the thoracic aorta was good.

Resting levels of blood flow in the arms and legs measured before and after operation by Slaughter and Wakim have shown no significant difference in the series of cases studied. Unfortunately there is no known method of making a good study of blood flow during exercise.

Howard B. Burchell, M.D.
Earl H. Wood, M.D.
Rochester, Minnesota

References

1. Burwell, C. S. and Dexter, Lewis: Venous Catheterization in Congenital Heart Disease. Mod. Concepts Cardiovasc. Dis. 16:2 pp. (Apr.) 1947.
2. Bing, R. J., Handelsman, J. C. and Campbell, J. A.: Physiologic Diagnostic Tests in Congenital Heart Disease. Mod. Concepts Cardiovasc. Dis. 17:2 pp. (Mar.) 1948.
3. Gibson, Stanley: Surgery for Congenital Cardiovascular Anomalies. Mod. Concepts Cardiovasc. Dis. 17:2 pp. (Apr.) 1948.
4. Taussig, Helen B.: Analysis of Malformations of the Heart Amenable to a Blalock-Taussig Operation. Am. Heart J. 36:321-333 (Sept.) 1948.
5. Hitzig, W. M.: The Value of Circulation Times. Mod. Concepts Cardiovasc. Dis. 16:2 pp. (Aug.) 1947.
6. Gross, R. E.: Surgical Treatment for Coarctation of the Aorta. Mod. Concepts Cardiovasc. Dis. 16:2 pp. (Feb.) 1947.

NOTES

NOTES